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【 CASE REPORT 】

Eagle Syndrome with Internal Carotid Artery Compression Causing Recurred Syncope

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Abstract:

We herein report a 54-year-old man with eagle syndrome who presented with repeated episodes of syncope, especially after moving his head to a downward position. Computed tomography with contrast revealed a bilateral elongated styloid process. The left internal carotid artery was obviously compressed by the left elongated styloid processes. A transcranial Doppler examination detected a significantly decreased blood flow velocity in the left middle cerebral artery when the patient slightly lowered his head position. After surgery, the positional cerebral blood flow alteration disappeared. No further similar syncope episodes have been reported to date.

Key words: Eagle syndrome, syncope, elongated styloid process, styloid-carotid syndrome, case report

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Introduction

Eagle syndrome (ES) is a relatively rare condition first described by Eagle in 1937 (1). It is characterized by a pathologically elongated styloid process or ossified stylohyoid ligament producing symptoms in the head and neck regions. Syncope is defined as the momentary loss of consciousness resulting from transient cerebral ischemia that can be caused by a variety of diseases, including alterations in the heart structure, lung disease, metabolic disorders, and epilepsy. In most cases, chronic painful neck swelling is the common complaint among patients presenting to the emergency department, but these cases can present diagnostic challenges.

We herein report an unusual case of ES with repeated episodes of syncope after changing the patient's head position.

Case Report

A 54-year-old right-handed man was admitted to our hospital due to several episodes of syncope. The first episode of syncope happened seven months ago, when the patient suffered from short-term loss of consciousness without any obvious inducement while watching over a chess competition in a standing posture. The second episode occurred when the patient was taking an elevator while glancing down at his feet. The third incident happened when the patient tried to lower his head after taking a shower to wipe his neck.

Before the onset of each of these syncope episodes, the patient had no general malaise, palpitations, sweating, blind eyes, limb weakness, or any other discomfort. A witness added that there were no remarkable symptoms, such as limb twitching, incontinence, tongue bite, or eyes rolling upward, before these syncope episodes. On average, the syncope lasted about three to five minutes, and the patient regained consciousness spontaneously after each occasion without any persistent discomfort. Since the first episode, the patient had complained of an unusual sensation of having a foreign body in the pharynx that could be touched in the submandibular region. However, after several rounds of consultation, doctors suggested that he not worry about it.

The patient denied having any remarkable medical or family history of ES but was a smoker. On admission, his vital signs were stable. On bimanual palpation, tender bony

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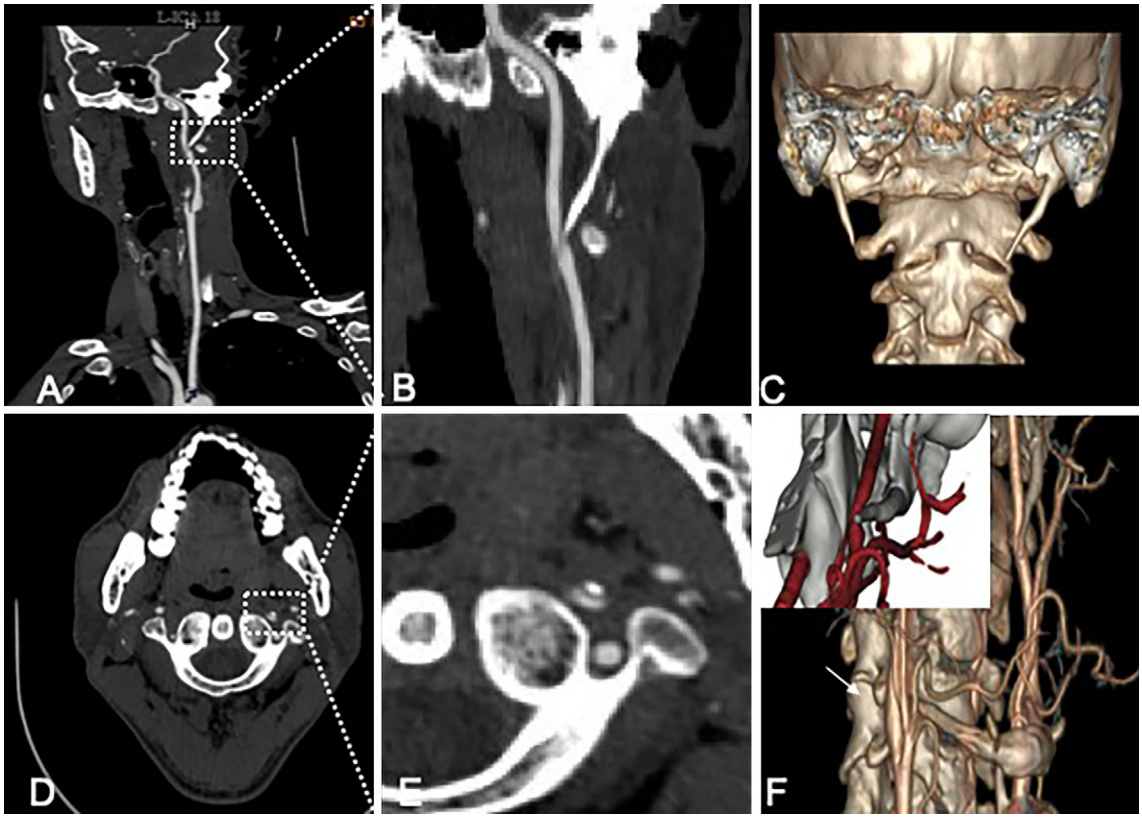

Figure 1. Contrast CT shows the relationship between the abnormally elongated styloid process and ICA. (A, B) CTA findings of the ICA and styloid process in the sagittal view. (C) 3-D reconstruction of CT images. The transverse view shows that the length of the right styloid process is 3.6 cm while that of the left is 4.4 cm. (D, E) CTA findings in the axial view reveal that the styloid process elongation has compressed the left ICA. (F) 3-D reconstruction of CTA images.

prominences in both tonsillar fossa could be touched. Other physical examination findings were unremarkable, except for left peripheral facial paralysis.

A systematic investigation was conducted to determine the pathogenesis of his syncope. The findings of laboratory examinations, including the leukocyte count, hemoglobin count, inflammatory factors, renal function, liver function, cardiac function, electrolyte level, D-dimer level, and thyroid function, were not significantly abnormal. 24-h Holter monitoring indicated sinus rhythm with an average heart rate of 78 beats/min without ST-T changes. A video electroencephalogram (EEG) showed unremarkable results.

Brain magnetic resonance imaging (MRI) revealed a normal brain structure. However, cervical computed tomography (CT) with contrast showed bilaterally elongated abnormal styloid processes that could be clearly visualized with three-dimensional (3-D) reconstruction. The lengths of the enlarged styloid processes were 4.4 cm on the left side and 3.6 cm on the right side. CT angiography (CTA) demonstrated obvious compression of the left internal carotid artery (ICA) by the ipsilateral elongated styloid processes (Fig. 1). Real-time transcranial Doppler (TCD) monitoring showed that the mean blood flow velocity of the left middle cerebral artery (MCA) was 236/105 cm/s, and the pulse index was normal when the patient was in the normal sitting position. The fast velocity of the MCA was caused by the beaded stenosis detected on CTA.

Notably, the bilateral blood flow velocity of the MCA did not change significantly when the patient turned his head to the left or right or tilted his head backward. However, immediately after tilting his head forward, the blood flow rate of the left MCA was significantly decreased, and the velocity was reduced to 80/43 cm/s. At the same time, the patient complained of sudden dizziness and other pre-syncope conditions. Interestingly, immediately after returning to the normal sitting position, the blood flow rate of the left MCA was gradually restored with simultaneous disappearance of the pre-syncope symptoms (Fig. 2). Based on the patient's medical history and analyses of his past and present clinical symptoms, he was diagnosed with ES or the abnormal elongation of styloid process-induced recurrent syncope.

After conducting detailed discussions with the patient and his family members, a transcervical styloidectomy approach was performed to manage the ES symptoms. No other treatment was prescribed to the patient during the postoperative or follow-up period. Postoperative TCD demonstrated no significant changes in the blood flow velocity of the left MCA when changing the position of his head, even after lowering his head (Fig. 3).

During one year of post-surgery follow-up, the patient

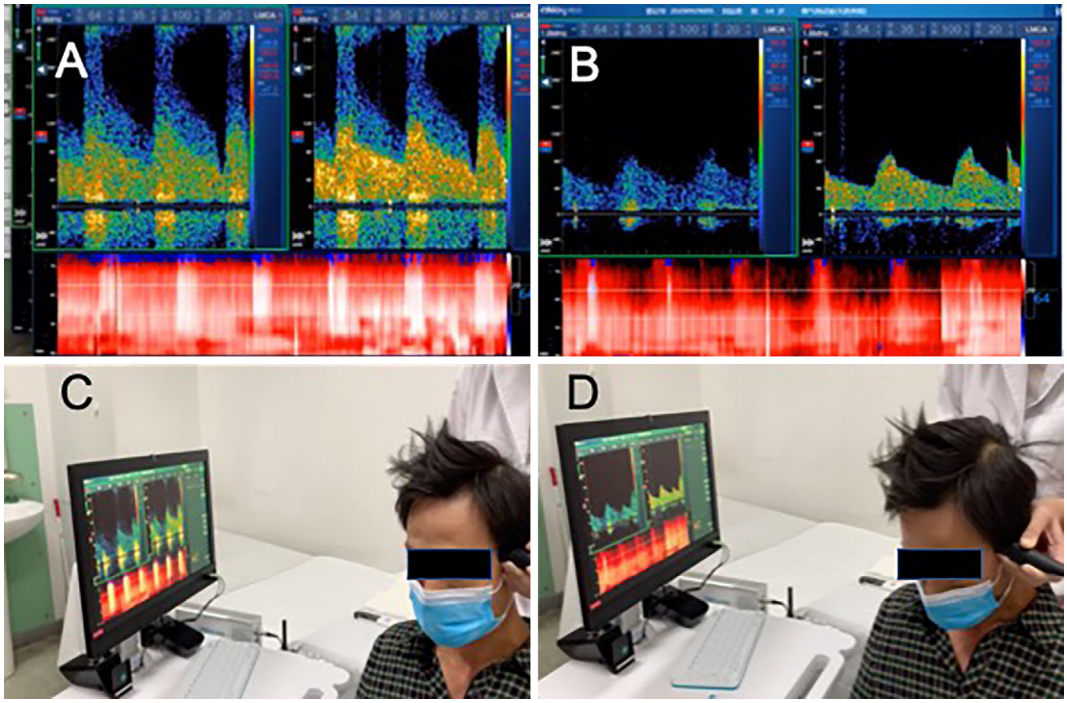

Figure 2. **Application of real-time TCD to monitor the blood flow changes in the left MCA. (A, C) The max blood flow velocity of the left MCA was 236/105 cm/s in the normal sitting position. However, the velocity significantly decreased significantly when the patient tried to look down, reporting an instantaneous feeling of dizziness (B, D).**

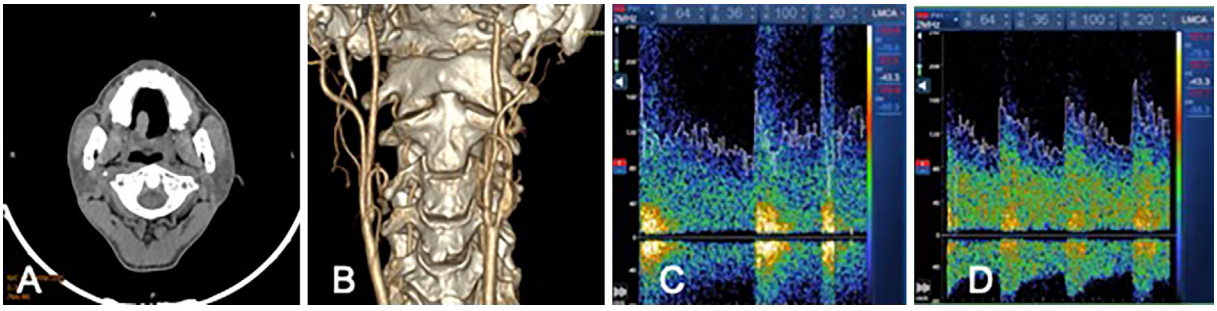

Figure 3. **Follow-up information. After surgery, (A) CTA in the sagittal view shows no compression of the LICA (B) and a shortened styloid process. (C, D) No significant change in the blood flow velocity of the LMCA is seen after turning or lowering his head.**

never experienced any further episodes of syncope or any other pre-syncope symptoms.

Discussion

The styloid process is a thin bony projection of the temporal bone extending inferiorly to its adjacent structures, including carotid arteries, internal jugular vein, cranial nerves VII, IX, X, and XII (2). The average length of the styloid process is 2.5-3 cm, and a length beyond 3 cm is considered abnormal elongation. An abnormal morphology, length, and orientation of the styloid process and/or ossification of the styloid hyoid ligament may result in corresponding clinical symptoms.

Two types of ES have been reported thus far: the classic form and the variant form. Clinical symptoms of classic ES include neck pain, pharynx pain, pharynx foreign body sensation, and tongue pain, with these symptoms commonly misdiagnosed as cervicofacial pain, such as trigeminal neuralgia, glossopharyngeal neuralgia, migraine, and even cervical arthritis (3). Our patient exhibited left peripheral facial paralysis, which might have been caused by local compression of the facial nerve. The variant form of ES is also known as vascular ES. Mechanical compression may cause stenosis in the adjacent arteries, especially in the ICA, resulting in transient cerebral ischemic events (4, 5). The distance between the ICA and the styloid process can be considered when assessing risk factors for carotid artery dissection (CAD). Indeed, patients with ES are reportedly four times more likely to have CAD than normal subjects (6).

Persistent inter-connection between the ES and ICA may lead to repetitive ICA micro-trauma, resulting in the pseu-

doaneurysm formation with internal thrombus (7, 8). Furthermore, the jugular vein can also be compressed by abnormal elongation of the styloid process and the lateral mass of the first cervical vertebral body (C1), leading to increased jugular pressure, insomnia, tinnitus, head noises, headache, earache, and other discomfort. ES is also a potential cause of idiopathic cranial hypertension (ICH) (9). In addition, it has been reported that glossopharyngeal neuralgia secondary to ES can lead to seizure attacks in elderly patients (10).

Considering the abundant collateral circulation in the brain, syncope caused by compression of the ipsilateral ICA due to a prolonged styloid is rare, and common symptoms include contralateral limb weakness, numbness, aphasia, and other manifestations due to ischemic events (11). In the present case, all syncope episodes occurred immediately after changing the position of his head, mostly in the downward direction, and TCD monitoring results indicated that the blood flow velocity of the left MCA was significantly reduced, along with the pulse index. We therefore considered that the most likely reason for the syncope was a rapid decrease in the intracranial blood flow due to the compression of the ICA by the extended process under the premise that the circle of Willis was not fully opened. The vasovagal reflex caused by extrinsic compression of the carotid plexus might also have been involved in the syncope.

Medical history and physical examination findings are critical in diagnosing ES, with CT considered the gold-standard modality for the diagnosis. The length, width, and angulation of the styloid process correlated with possible symptoms can be clearly displayed by CT and 3-D reconstruction of CT images. CTA can be used to evaluate the relationship with surrounding vessels, thereby avoiding further digital subtraction angiography (DSA) (12, 13). TCD is an effective and non-invasive tool for both the diagnosis and treatment of ES patients. To better understand the 'dynamic' relationship between the styloid process and surrounding vascular structure, clinicians usually suggest performing TCD in the same five standardized positions of the head: rest position, maximum extension, maximum flexion of the head, maximum right, and maximum left rotation (14).

Management of ES varies depending on the patient's symptoms, as this condition can be treated either conservatively or surgically or both. Conservative therapeutic approaches typically include administration of non-steroidal anti-inflammatory drugs (NSAIDs), a combination of anticonvulsants, antidepressants, and local injections of steroid hormones to manage the pain. For vascular variant ES, antiplatelet medications, such as aspirin, are considered reasonable treatment options for transient ischemic stroke, while short-term anticoagulation approaches have been suggested to treat CAD. Whether or not the surgical removal of the long styloid process constitutes a general treatment for ES has not yet been clarified. Some authors consider stenting or surgical removal to treat CAD as being potentially harmful or even life-threatening, carrying the possibility of stent fracture as previously reported (15). For patients with jugular vein compression, microsurgical decompression of the jugular vein has been found effective (16, 17). The safety and effectiveness of removing the stenosis bone to improve cranial hypertension still need to be closely examined in a larger number of cases (18). The majority of patients show significant symptom improvement after surgery, especially for neck or joint pain. However, while surgical excision can relieve the pain, the improvement of headache symptoms after styloidectomy is not very significant (19). In the present patient, styloidectomy was performed by a skilled team of otolaryngologists, and the patient has had no more episodes of syncope to date.

Conclusion

When dealing with cases of unexplained syncope, we suggest that ES be considered as a differential diagnosis, especially when the syncope happens in patients due to specific postures or neck movements.

The authors state that they have no Conflict of Interest (COI).

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